

CASE REPORT

Double-lumen Aortic Arch: Persistence of the Fifth Aortic Arch?

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Abstract

Double-lumen aortic arch is a rare congenital anomaly related to persistence of the fifth aortic arch. It may be found alone or in association with other anatomical changes of the heart. We report a case of double-lumen aortic arch associated with coarctation of the aorta and patent ductus arteriosus in a child with a congenital malformation known as the VACTERL association (vertebral defects, imperforate anus, cardiopathy, tracheoesophageal fistula, renal abnormalities and limb anomalies).

Introduction

Double-lumen aortic arch is a rare, usually underdiagnosed congenital anomaly, possibly secondary to the persistence of the fifth embryonic aortic arch or the presence of dorsal collateral arteries that connect the fourth and sixth aortic arch.¹⁻³ The authors report a case of double-lumen aortic arch associated with coarctation of the aorta and ductus arteriosus in a child with congenital malformation characterized by presence of vertebral defects, imperforate anus, heart disease, tracheoesophageal fistula, renal anomalies and limb abnormalities, known as the VACTERL association, which was accurately diagnosed by echocardiography and confirmed by computed tomography.

Keywords

Aorta, Thoracic/ abnormalities; Vascular Ring; Echocardiography/ methods; Vascular Malformation/ diagnostic imaging.

Case Report

Seven-year-old child with VACTERL association, with previous history of surgical treatment for ductus arteriosus and repair of aortic coarctation in the first year of life. During hospitalization for reconstruction of intestinal transit, an echocardiography revealed dextrocardia, bicuspid aortic valve, significant ascending aortic enlargement (z-score +5.04) and transverse aorta with two lumens, compatible with double-lumen aortic arch. Both anterior and posterior aortic lumens were unobstructed, with laminar flow, measuring about 14.6 mm and 6.5 mm, respectively. The cephalic veins emerged from the anterior lumen, and the posterior lumen originated proximal to the brachiocephalic artery and ended in the descending aorta (Figures 1a and b, 2a, video 1). The echocardiographic findings were confirmed by chest computed tomography, which also showed the presence of two brachiocephalic trunks (Figure 2b).

Discussion

Double-lumen aortic arch, an exceptionally rare congenital anomaly,⁴ was first described in humans by Van Praagh and Van Praagh⁵ in 1969, and persistence of the fifth aortic arch was proposed as the cause of this condition. Descriptions of persistence of the fifth arch in the literature have been limited to case reports. Brown in 1913 and Huntington in 1919 reported the first hypotheses for the development of this persistent arch.⁴

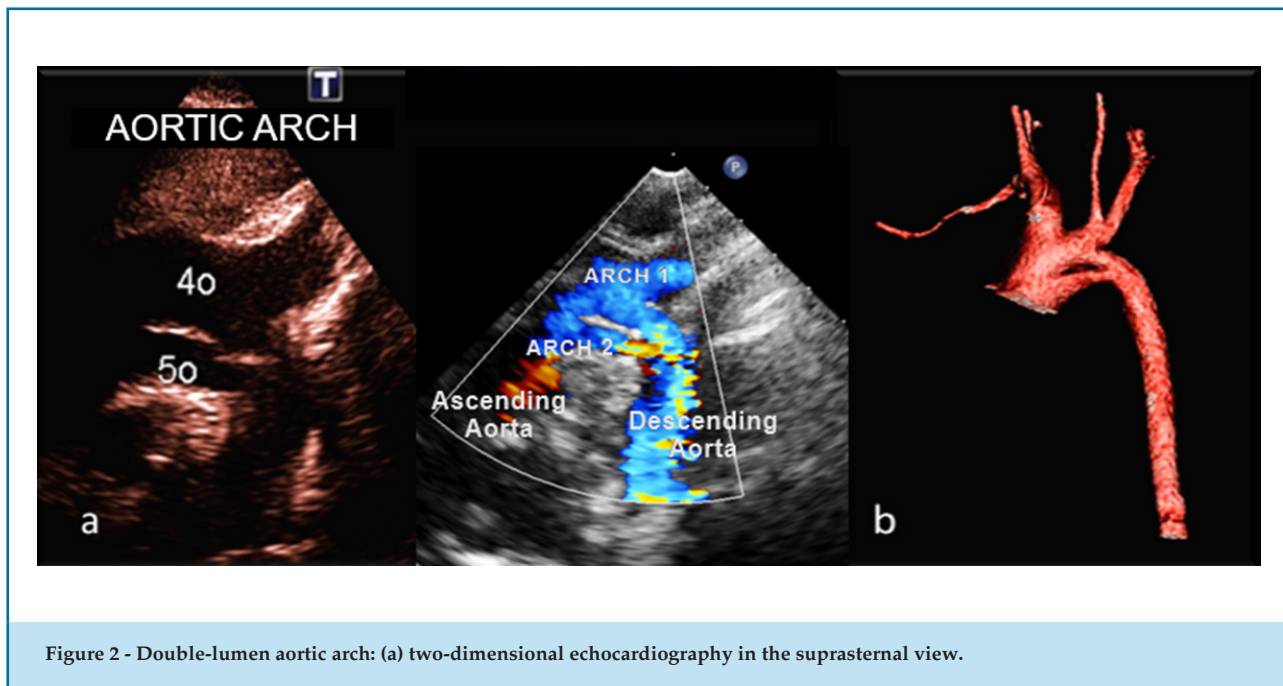
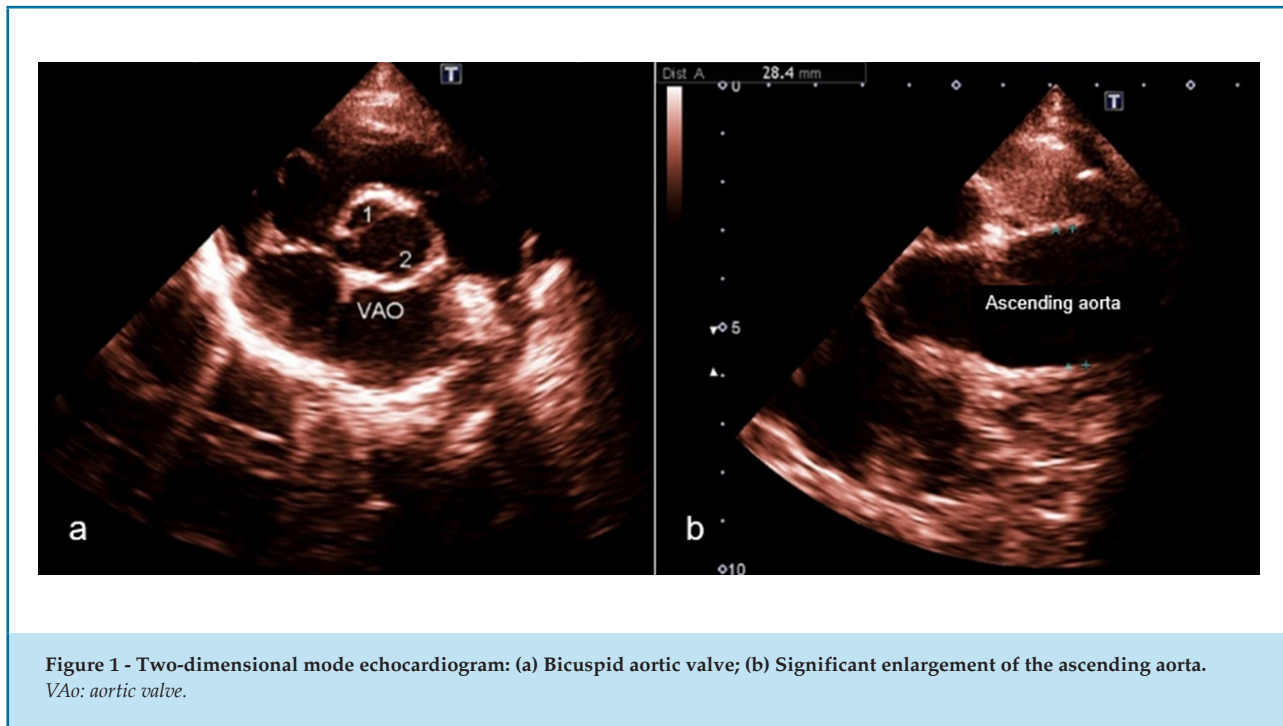
However, Gupta et al.,² studying developing human embryos, identified remnants of an unambiguous fifth arch artery in only one embryo and showed that dorsal collateral canals connecting the fourth and sixth arches are much more common and may best explain the formation of a double aortic lumen. According to these authors, a persistent fifth aortic arch is defined as an

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extrapericardial arch that arises from the ascending aorta proximal to the brachiocephalic artery, runs a serpentine course and terminates in the dorsal aorta or in the sixth aortic arch of the pulmonary artery.²

Double-lumen aortic arch can be classified into:

- I. Double lumen with both lumens patent;
- II. Atresia or interruption of one (upper) lumen and patency of the other;
- III. Connection between the lower lumen and the pulmonary artery.^{6,7}

Although frequently associated with several cardiac defects, such as coarctation or interruption of the aorta, patent ductus arteriosus, tetralogy of Fallot, atrioventricular septal defect, truncus arteriosus, pulmonary and tricuspid atresia, this anomaly can also be found alone.^{3,8}

The clinical presentation of double-lumen aortic arch depends on the type of connection and presence of associated anomalies. Type I, as reported here, is the most common form, usually of no clinical significance. Types II and III have positive hemodynamic consequences, by providing an alternative systemic arch in case of coarctation or aortic arch interruption (type II), and by acting as a systemic-pulmonary shunt when associated with pulmonary or tricuspid atresia (type III).^{3,6-8}

In the present case, both arches ran on the same side of the trachea and, unlike the classical double aortic arch, which involves both trachea and esophagus, and does not result in a vascular ring.⁶ Thus, it may correspond to a non-classical double arch possibly secondary to the presence of collaterals between the fourth and sixth artery arches, as described by Gupta et.al. Differential diagnosis should be made with patent ductus arteriosus and aortopulmonary window, especially in type III.^{1,3,6,7}

Echocardiography allows the detection of two lumens of the aortic arch running in parallel and helps in the identification of coexisting anomalies. Computed tomography and magnetic resonance imaging help define the anatomy of the aortic arch and its variants, confirm the diagnosis of this anomaly and detect possible coexisting abnormalities such as pulmonary or cardiovascular diseases.⁶

Conclusion

Type I aortic double lumen is an occasional finding with no significant hemodynamic consequences. We

described a case of double-lumen aortic arch, possibly caused by factors related to a non-classical double arch, including formation of dorsal collaterals, rather than a persistence of the fifth aortic arch. Detection of this anomaly is important for familiarization with the various presentations of the aortic arch.

Author contributions

Conception and design of the research: Monteze NM, Guimaraes AFM. Acquisition of data: Monteze NM, Guimaraes AFM. Analysis and interpretation of the data: Monteze NM, Guimaraes AFM, Araujo FDR. Writing of the manuscript: Monteze NM. Critical revision of the manuscript for intellectual content: Monteze NM, Guimaraes AFM, Araujo FDR.

Potential Conflict of Interest

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Study Association

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Ethics approval and consent to participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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